Complete Summary

GUIDELINE TITLE

Cirrhosis.

BIBLIOGRAPHIC SOURCE(S)

Cirrhosis. Philadelphia (PA): Intracorp; 2005. Various p. [20 references]

GUIDELINE STATUS

This is the current release of the guideline.

All Intracorp guidelines are reviewed annually and updated as necessary, but no less frequently than every 2 years. This guideline is effective from April 1, 2005 to April 1, 2007.

COMPLETE SUMMARY CONTENT

SCOPE

DISEASE/CONDITION(S)

Cirrhosis

GUIDELINE CATEGORY

Diagnosis
Evaluation
Management
Treatment

CLINICAL SPECIALTY

Family Practice
Gastroenterology
Internal Medicine
Surgery

INTENDED USERS

Allied Health Personnel
Health Care Providers
Health Plans
Hospitals
Managed Care Organizations
Utilization Management

GUIDELINE OBJECTIVE(S)

To present recommendations for the diagnosis, treatment, and management of cirrhosis that will assist medical management leaders to make appropriate benefit coverage determinations

TARGET POPULATION

Individuals with cirrhosis

INTERVENTIONS AND PRACTICES CONSIDERED

Diagnosis/Evaluation

1. Physical examination and assessment of signs and symptoms
2. Diagnostic tests:
    - Laboratory studies
        - Chemistry
        - Renal function
        - Liver enzymes
        - Other blood work
    - Ultrasonography
    - Computed tomography (CT)
    - Technetium-99m sulfur colloid scanning
    - Percutaneous liver biopsy
    - Diagnostic paracentesis
    - Endoscopic gastroduodenoscopy

Management/Treatment

1. Restricted exposure to hepatic toxins (e.g., alcohol, acetaminophen)
2. Sodium and fluid restriction
3. Iron and folic acid supplement
4. Vitamin K, fresh frozen plasma
5. Blood transfusion
6. Lactulose
7. Medications: diuretics, beta-blockers, antibiotics

8. Endoscopy with sclerosis
9. Transjugular intrahepatic portosystemic stent shunt procedure (TIPSS)
10. Referral to specialists
11. Liver transplantation

MAJOR OUTCOMES CONSIDERED

Utility of diagnostic tests

METHODOLOGY

METHODS USED TO COLLECT/SELECT EVIDENCE

Hand-searches of Published Literature (Primary Sources)
Hand-searches of Published Literature (Secondary Sources)
Searches of Electronic Databases

DESCRIPTION OF METHODS USED TO COLLECT/SELECT THE EVIDENCE

Searches were performed of the following resources: reviews by independent medical technology assessment vendors (such as the Cochrane Library, HAYES); PubMed; MD Consult; the Centers for Disease Control and Prevention (CDC); the U.S. Food and Drug Administration (FDA); professional society position statements and recommended guidelines; peer reviewed medical and technology publications and journals; medical journals by specialty; National Library of Medicine; Agency for Healthcare Research and Quality; Centers for Medicare and Medicaid Services; and Federal and State Jurisdictional mandates.

NUMBER OF SOURCE DOCUMENTS

Not stated

METHODS USED TO ASSESS THE QUALITY AND STRENGTH OF THE EVIDENCE

Weighting According to a Rating Scheme (Scheme Not Given)

RATING SCHEME FOR THE STRENGTH OF THE EVIDENCE

Not stated

METHODS USED TO ANALYZE THE EVIDENCE

Review

DESCRIPTION OF THE METHODS USED TO ANALYZE THE EVIDENCE

Not stated

METHODS USED TO FORMULATE THE RECOMMENDATIONS

Expert Consensus (Delphi)

DESCRIPTION OF METHODS USED TO FORMULATE THE RECOMMENDATIONS

A draft Clinical Resource Tool (CRT or guideline) is prepared by a primary researcher and presented to the Medical Technology Assessment Committee or the Intracorp Guideline Quality Committee, dependent upon guideline product type.

The Medical Technology Assessment Committee is the governing body for the assessment of emerging and evolving technology. This Committee is comprised of a Medical Technology Assessment Medical Director, the Benefit and Coverage Medical Director, CIGNA Pharmacy, physicians from across the enterprise, the Clinical Resource Unit staff, Legal Department, Operations, and Quality. The Intracorp Guideline Quality Committee is similarly staffed by Senior and Associate Disability Medical Directors.

Revisions are suggested and considered. A vote is taken for acceptance or denial of the CRT.

RATING SCHEME FOR THE STRENGTH OF THE RECOMMENDATIONS

Not applicable

COST ANALYSIS

A formal cost analysis was not performed and published cost analyses were not reviewed.

METHOD OF GUIDELINE VALIDATION

Comparison with Guidelines from Other Groups
Internal Peer Review

DESCRIPTION OF METHOD OF GUIDELINE VALIDATION

Not stated

RECOMMENDATIONS

MAJOR RECOMMENDATIONS

Diagnostic Confirmation

Subjective Findings

- History of blood transfusion, intravenous (IV) drug abuse (hepatitis)
- History of alcohol abuse
- History of exposure to hepatotoxic agents

- Family history of liver disease (hemochromatosis, a-1 antitrypsin deficiency)
- Fatigue
- Report of abdominal pain in the right upper quadrant (RUQ)
- Dyspepsia
- Pruritus
- Shortness of breath
- Multiple infections
- Unintentional weight loss
- Patient may be asymptomatic

Objective Findings

- Skin
    - Jaundice
    - Pallor
    - Caput medusa (dilated superficial periumbilical vein)
    - Palmar erythema
    - Spider angiomata
    - Ecchymosis (thrombocytopenia or coagulation factor deficiency)
    - Hyperpigmentation (hemochromatosis)
    - Scabbed, scratched areas
- Eyes
    - Scleral icterus (yellowed whites)
    - Kayser-Fleischer corneal rings
        - Wilson's disease - caused by copper deposits
        - Best seen on slit-lamp examination
- Head and Neck
    - Fetor hepaticus (musky/sweet and sour breath)
    - Parotid or lacrimal gland enlargement
- Central Nervous System
    - Flapping or "pill-rolling" tremor
    - Asterixis (hepatic encephalopathy)
    - Choreoathetosis
    - Dysarthria (Wilson's disease)
    - Confusion
    - Peripheral neuropathy
- Abdomen
    - Tender hepatomegaly (congestive hepatomegaly)
    - Small, nodular liver (cirrhosis)
    - Spontaneous bacterial peritonitis (primary biliary disease)
    - Palpable spleen (portal hypertension)
    - Ascites (portal hypertension, hypoalbuminemia)
    - Venous hum auscultated over periumbilical veins (portal hypertension)
    - Gastrointestinal or variceal bleeding
- Genitourinary
    - Bilirubinurea
    - Hirsutism
    - Hypogonadism
        - Gynecomastia or altered body hair pattern in males
        - Testicular atrophy (hemochromatosis)
        - Erectile dysfunction
        - Oligomenorrhea, amenorrhea, and sterility in women

- Extremities
    - Dupuytren's contracture
    - Pedal edema (hypoalbuminemia, right-sided heart failure)
    - Arthropathy
- General
    - Cachexia
    - Other signs of nutritional compromise
    - Deconditioned status; activities of daily living (ADL) compromised

Diagnostic Tests

- Laboratory Studies
    - Chemistry
        - Hypokalemia - low serum potassium (K+)
        - Hyponatremia - low serum sodium (Na+)
        - Serum ammonia elevated (NH4)
        - Serum copper elevated (Cu++) - Wilson's disease
    - Renal function
        - Creatinine increased
        - Blood urea nitrogen (BUN) increased
    - Liver enzymes
        - Aspartate transaminase (AST) elevated
        - Alanine transaminase (ALT) elevated
        - Alkaline phosphatase elevated
    - Other blood work
        - Bilirubin elevated
        - Albumin low
        - Prothrombin time (PT) prolonged
        - Ferritin abnormally low
        - Total iron binding capacity (TIBC) also low
        - Hemoglobin/hematocrit low
        - Antinuclear antibodies (ANA) may be found in autoimmune hepatitis
        - (+) Hepatitis viral titers
- Ultrasonography
    - Of limited use diagnostically
    - Abnormal findings in advanced disease
    - Primary usefulness for evaluating vascular patency, gallstone detection, identifying masses or bile-duct dilation/obstruction
- Computed tomography (CT) scan
    - Of limited use diagnostically
    - Abnormal findings in advanced disease
    - Detects mass lesions in liver, pancreas
    - Valuable to assess fat content in liver
    - Also identifying idiopathic hemochromatosis, Budd-Chiari syndrome
    - Useful to detect dilation of intrahepatic bile ducts, varices, splenomegaly
- Technetium-99m sulfur colloid scanning
    - Cirrhosis identified by shift of colloid uptake to the spleen, bone marrow
    - Hepatic adenoma identified by "cold" defect

    - Budd-Chiari syndrome identified by increased uptake in the caudate lobe
- Percutaneous liver biopsy - useful in diagnosis if questionable etiology
- Diagnostic paracentesis for new-onset ascites
- Endoscopic gastroduodenoscopy (EGD)
    - Confirms if varices present, detection of specific causes if gastrointestinal (GI) bleeding

Differential Diagnosis

- Hepatitis - viral, drug induced, or autoimmune (see the Intracorp guideline Hepatitis)
- Hepatocellular carcinoma
- Wilson's disease
- Primary sclerosing cholangitis
- Cholangiocarcinoma
- Biliary duct disease - cholangitis or stone disease
- Pancreatitis (see the Intracorp guideline Pancreatitis)
- Pancreatic cancer (see the Intracorp guideline Pancreatic Cancer)
- Sarcoiditis
- Right-sided heart failure
- Constrictive pericarditis (see the Intracorp guideline Pericarditis)
- Pulmonary hypertension
- Idiopathic portal hypertension
- Budd-Chiari syndrome, also known as hepatic vein thrombosis
- Veno-occlusive disease
- Myeloid metaplasia

Treatment

Treatment Options

- Restrict exposure to any hepatic toxins (e.g., alcohol, acetaminophen)
- Dietary restrictions as well as supplements
    - Sodium and fluid restriction; diuretics for ascites and edema
    - Iron and folic acid supplements for anemia
- For hemorrhagic tendencies
    - Vitamin K, fresh frozen plasma
    - Blood transfusion (packed red blood cells [PRBCs]) may be needed
- Lactulose for hepatic encephalopathy
- Antibiotics for ascites to prevent spontaneous bacterial peritonitis (SBP)
- Beta-blockade (beta-blocker medication) for portal hypertension
- Endoscopy with sclerosis for variceal bleeds
- Transjugular intrahepatic portosystemic stent shunt procedure (TIPSS)
- Liver transplantation - orthotopic liver transplant (OLT)

Duration of Medical Treatment

- Medical
    - Cirrhosis is a chronic disease, and treatment will be ongoing
- Surgical

- Appropriate candidates for liver transplant will require life-long follow up

Additional information regarding primary care visit schedules, referral options, specialty care, and durable medical equipment is provided in the original guideline document.

The original guideline document also provides a list of red flags that may affect disability duration, and return to work goals, including

- Fatty changes to liver
- Severe disruption of liver functions

CLINICAL ALGORITHM(S)

None provided

EVIDENCE SUPPORTING THE RECOMMENDATIONS

TYPE OF EVIDENCE SUPPORTING THE RECOMMENDATIONS

The type of supporting evidence is not specifically stated for each recommendation.

BENEFITS/HARMS OF IMPLEMENTING THE GUIDELINE RECOMMENDATIONS

POTENTIAL BENEFITS

Appropriate diagnosis, treatment, and management of cirrhosis that assist medical management leaders to make appropriate benefit coverage determinations

POTENTIAL HARMS

Not stated

IMPLEMENTATION OF THE GUIDELINE

DESCRIPTION OF IMPLEMENTATION STRATEGY

An implementation strategy was not provided.

INSTITUTE OF MEDICINE (IOM) NATIONAL HEALTHCARE QUALITY REPORT CATEGORIES

IOM CARE NEED

Living with Illness

IOM DOMAIN

Effectiveness

IDENTIFYING INFORMATION AND AVAILABILITY

BIBLIOGRAPHIC SOURCE(S)

Cirrhosis. Philadelphia (PA): Intracorp; 2005. Various p. [20 references]

ADAPTATION

Not applicable: The guideline was not adapted from another source.

DATE RELEASED

1997 (revised 2005)

GUIDELINE DEVELOPER(S)

Intracorp - Public For Profit Organization

SOURCE(S) OF FUNDING

Intracorp

GUIDELINE COMMITTEE

CIGNA Clinical Resources Unit (CRU)
Intracorp Disability Clinical Advisory Team (DCAT)
Medical Technology Assessment Committee (MTAC)
Intracorp Guideline Quality Committee

COMPOSITION OF GROUP THAT AUTHORED THE GUIDELINE

Not stated

FINANCIAL DISCLOSURES/CONFLICTS OF INTEREST

Not stated

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AVAILABILITY OF COMPANION DOCUMENTS

The following is available:

- Policies and procedures. Medical Technology Assessment Committee Review Process. Philadelphia (PA): Intracorp; 2004. 4 p.
- Online guideline user trial. Register for Claims Toolbox access at www.intracorp.com.

Licensing information and pricing: Available from Intracorp, 1601 Chestnut Street, TL-09C, Philadelphia, PA 19192; e-mail: lbowman@mail.intracorp.com.

PATIENT RESOURCES

None available

NGC STATUS

This NGC summary was completed by ECRI on May 31, 2005. The information was verified by the guideline developer on June 7, 2005.

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